**Scholars Journal of Medical Case Reports** **ISSN 2347-6559 (Online)**
Sch J Med Case Rep 2014; 2(6):424-426 **ISSN 2347-9507 (Print)**

(An International Publisher for Academic and Scientific Resources) DOI: 10.36347/sjmcr.2014.v02i06.023

# Ossifying Fibroma Presenting As a Nasal Mass: A Diagnostic Dilemma

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**Abstract:** Ossifying fibroma (OF) is a benign, slow-growing, central bone tumor, usually of the jaws, especially the mandible, composed of fibrous connective tissue within which bone is formed. The overlapping clinical and histopathological features of these subtypes have led to diagnostic dilemma and confusion. Complete excision of this tumor has become a necessity since it is notorious for recurrence. Here is a case of two month old child diagnosed to have ossifying fibroma of nose. We report this case due to uncommon site of involvement and age group of the patient imparting uniqueness to this case.
**Keywords:** Ossifying Fibroma, Nose, Two month child, Juvenile age group, Fibroma, Nasal mass

## INTRODUCTION

Fibro-osseous lesions of sino nasal tract include a variety of neoplastic and non-neoplastic entities. The non neoplastic group includes fibrous dysplasia, aneurysmal bone cyst, giant cell granuloma. Included in neoplastic category are ossifying fibroma, giant cell tumor, fibromyxoma, osteoblastoma, osteosarcoma [1].

Ossifying fibroma (OF) is a rare benign fibro-osseous lesion which was first described by Menzel in 1872. He considered it as a form of Osteoma but the term of "Ossifying Fibroma" was subsequently coined by Montgomery in 1927 [1].

It is a destructive, deforming, slow growing, benign fibro-osseous tumor that can occur almost anywhere in the facial skeleton [2]. It is very closely related to other lesions such as fibrous dysplasia, periapical dysplasia [3]. This uncommon tumor can present a diagnostic dilemma for the clinician and the pathologist, owing to overlapping clinical and histomorphologic features. The tumor can produce sinus obstruction, infection, facial deformity, proptosis and intracranial complications, even though it can remain asymptomatic in the early stage. Therefore the tumor needs to be excised completely in order to prevent recurrence [2].

A subset of craniofacial ossifying fibromas has been described with a predilection for the sino nasal tract. These usually occur in the first and the second decade of life [1]. However, a big ossifying fibroma requiring surgery at the age of two month has not been reported to the best of our knowledge. Here we report a case of a child presenting with a growth in the nose extending into the nasophaynx which on histopathological evaluation was diagnosed as ossifying fibroma. It was surgically removed and the child is doing well during post operative follow up.

## CASE REPORT

A two month old female child was brought to the ENT OPD with the chief complaints of inability to sleep in the supine position, for which he preferred to lie in the semi prone position, associated with noisy breathing and gasping for breath while taking breast feed, since 4 months. The child was asymptomatic and growing well prior to this period. From the onset, she had no vertigo, apparent nasal obstruction, nasal mucus, epistaxis, hyposmia or headache. Furthermore, she denied any history of trauma to the head or face.

ENT examination in the OPD revealed blocked nasal passage with no air entry and the child was a mouth breather. Anterior rhinoscopy revealed a growth. Examination of oral cavity revealed an inferiorly pushed hard and soft palate with no growth in the orophaynx. Posterior rhinoscopy could not be done. CT scan of PNS and Nasophaynx was earned out followed by examination in OT with biopsy of lesion. The CT scan revealed a big soft tissue mass with minimal enhancement and multiple areas of necrosis in the left nose and extending to the nasophaynx. The tumor did not cause any bone erosion and was not attached to the base of skull. There was no breach in the skull base. Definite site of origin could not be identified but the dense necrosis pattern in the nasal part pointed towards

a primary nasal tumor extending into the nasopharynx. The histopathological evaluation of the specimen included the presence of trabeculae of smaller bone, a variable amount of vascularised fibrous stoma and osteoblastic rimming of the trabeculae, qualifying it as Ossifying fibroma (Fig. 1, 2).

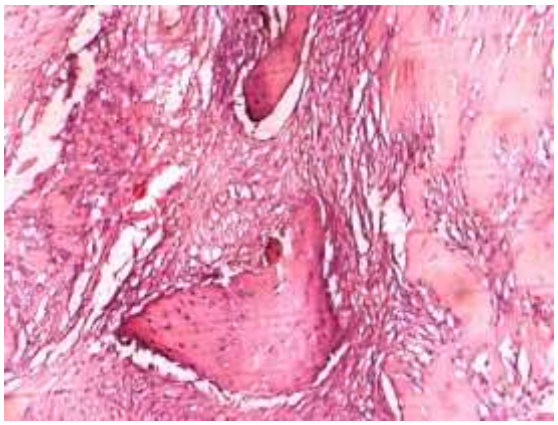

**Fig 1: H&E view of ossifying fibroma (H&E, 100x)**

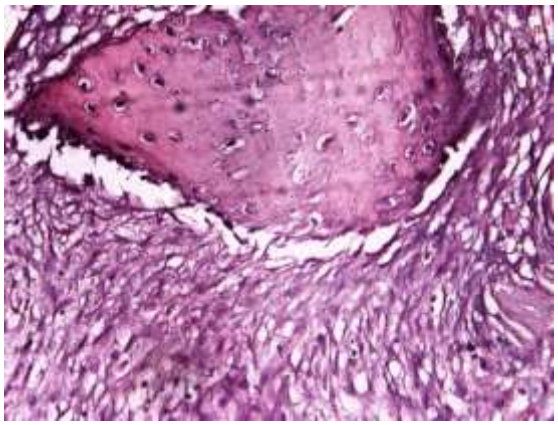

**Fig 2: H&E view of ossifying fibroma (H&E, 200x)**

Elective surgical excision was planned and during surgery it was found that the tumor was attached the bony septum on the left side. Hence it was inferred that this was the primary site of origin. The child is under follow up and is doing fine.

## DISCUSSION

Ossifying fibroma as reported in literature is commonly found in the tibia and fibula of children 10 years or younger but young adult individuals with an average age of 20 to 30 years are the most commonly affected groups with involvement of the head & neck region. In this site, the lesions arise in the mandible in about 62% to 89% of patients followed by the maxilla and rarely the orbit, skull base and calvarium. Women are affected more often than men with a female to male ratio of 2:1. Ossifying fibroma of the sinonasal tract occurs at a slightly older age ($3^{rd}$ to $4^{th}$ decade of life), and preferentially in black women. There is no evidence of hereditary predominance [1]. But our patient is a two month old child with nasal mass.

Since the first case of ossifying fibroma was reported in 1872, there was a period of time when ossifying fibroma and fibrous dysplasia of bone were considered to be the same disease. Following a long period of observation and study, they are now considered to be two different diseases. Ossifying fibroma is a bone tissue-derived benign tumor, whereas fibrous dysplasia is a hyperplastic bone lesion caused by bone mesenchymal dysplasia [5].

The clinical presentation of these tumors is variable, depending on the site and rate of growth. It ranges from an asymptomatic bone lesion found incidentally on imaging taken for another reason as in our case, to symptoms due to mass effect of sinonasal lesions such as nasal obstruction, anosmia, hyposmia, headache or epistaxis. Ocular symptoms include visual loss, diplopia, proptosis and epiphora. Larger tumors may also lead to a painless swelling of the involved bone. While pain and paresthesia are rarely associated with an ossifying fibroma where the temporal bone is involved, the patient may complain of pain, pulsatile tinnitus, otorrhea with progressive hearing loss. Meningitis and pneumocephalus are two rare intracranial complications of ossifying fibroma [1].

Histologically, the ossifying fibromas are well circumscribed, occasionally encapsulated, consisting of cellular fibrous tissues and thin isolated trabeculae of bones. The bone may show osteoblastic rimming and spherical deposits of calcified material, which are relatively acellular resembling cementum. The lack of consistent osteoblastic rimming of the bone trabeculae in fibrous dysplasia is used to distinguish it from an ossifying fibroma, which is more commonly rimmed by plump osteoblasts. Most authors consider fibrous dysplasia and ossifying fibroma to be histologically similar—with the sole differentiating feature being a fibrous capsule surrounding the latter and infrequently observed in the case of fibrous dysplasia. However, aggressive form of ossifying fibroma may lose its fibrous capsule.

Differential diagnoses include fibrous dysplasia, sinonasal psammomatous meningioma, and well-differentiated osteosarcoma. Fibrous dysplasia (FD) is an idiopathic non-neoplastic disease affecting patients during the first 2 decades of life. Craniofacial involvement is seen in 50% of patients with polyostotic lesion and 25% with monostotic lesions. Radiographically, FD tends to have more diffused margins. Clinically, FD displays self-limited growth with skeletal maturation and often reveals cessation of growth once adulthood is reached. The histological features include irregular shaped trabeculae of osteoid

or woven bone ("Chinese letter" configuration) dispersed in a fibrous stroma with variable cellularity, which is directly fused to peripheral normal bone. FD is often polyostotic whereas ossifying fibroma lesions are monostotic. The fibrous stroma in FD is often less vascular and cellular than OF without osteoblastic rims, but OF usually presents it. In contrast to OF and other fibroosseous lesion, FD typically demonstrate a rather monotonous pattern throughout the lesions.

Psammomatous meningioma is not a true fibro-osseous lesion and it may show no identifiable connection with the central nervous system (primary/ectopic) or may extend from the central nervous system (secondary). They appear as polypoid masses that cause nasal obstruction.

Meningioma is distinguished from OF by the presence of whorls of meningothelial spindle cells; frequently with empty-appearing nuclei. Psammoma bodies lack the osteoblastic rimming seen in the calcified ossicle of Ossifying fibroma. The diagnosis of meningioma can be confirmed by dual immunoreactivity of the meningothelial cells for epithelial membrane antigen (EMA) and the mesenchymal marker vimentin. The stromal cells in OF and FD may be vimentin positive but lack immunoreactivity for EMA. Craniofacial osteosarcomas are destructive, poorly defined, osteolytic, osteosclerotic or mixed lesions. Anaplastic tumor cells are seen histopathologically admixed with areas of osteoid formation. They are aggressive tumors that are prone to local recurrence and distant metastasis [1].

If the lesions are small, they are treated by enucleation. However, larger lesions require radical resection. Recurrence rates of these aggressive forms of ossifying fibromas are about 30% to 38%. Thus a regular followup is necessary [6].

Ossifying fibroma is a benign fibro-osseous tumor of the craniofacial region that is diagnosed with a combination of clinical, radiological and pathological criteria. Due to the possibility of the presence of hybrid lesions in this tumor, it is preferable to remove it in mass and take multiple sections for histopathological reporting. This would avoid missing a particular subtype of the tumor that might need a different surgical management. The surgical approaches and techniques have also not been well defined especially in the young patients. Complete surgical excision of the tumor is possible when surgery is based on preplanned criteria.

Many synonymous nomenclatures exist for a single entity and the controversy in classification and staging of the subtypes in the literature has added to the confusion. Hence there is a need to highlight the points of controversy existing for this tumor so that they could be avoided through a consensus in future.

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